Alveolar hydatid disease of the brain

Report of four cases

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Four cases of alveolar hydatid disease of the brain encountered within 27 months in eastern Turkey are
reported. All of the patients were male farmers who presented with signs of cerebral tumor. Two of the patients
were shown to harbor hepatic lesions and one of them had pulmonary metastases. The cerebral lesions were
removed in toto and neurological recovery was obtained in all four patients. A review of the literature revealed
only five previously reported cases treated surgically. It is concluded that cerebral Echinococcus multilocularis
lesions are amenable to surgery, and that their removal provides useful prolongation of life despite the presence
of hepatic or pulmonary disease.

KEY WORDS • alveolar hydatid disease • cerebral echinococcosis •
Echinococcus alveolaris • Echinococcus multilocularis

NEUROSURGEONS are rarely called upon to treat
alveolar hydatid disease. One of the most dan-
gerous of human infections, alveolar hydatid
disease is caused by Echinococcus multilocularis, also
known as Echinococcus alveolaris. This organism is a
parasite known in northern regions including Alaska,
Canada, Siberia, and the Alps, \(1,2\) but it has recently
been increasingly observed in the more temperate re-
gions of eastern France, Nebraska, and Illinois. \(2,22\) Un-
like Echinococcus granulosus, which produces cystic
lesions, \(E.\) multilocularis produces invasive lesions of
firm consistency, full of connective tissue and a jelly-
like material. The favorite location of these destructive
cysts is the liver; cerebral localization is rare.

We report four cases of alveolar hydatid disease of
the brain that were surgically treated at Atatürk Uni-
versity Medical School in Erzurum, Turkey, during a
period of 27 months. In each case, the cerebral lesion
was removed in toto and remission of neurological
deficits was obtained. The purpose of this paper is to
describe the clinical picture and surgical therapy of this
infestation and to alert neurosurgeons and pathologists,
particularly those in regions south of known endemic
areas of \(E.\) multilocularis, to the possibility of encoun-
tering such lesions. Cerebral \(E.\) multilocularis lesions
lend themselves to total removal with useful prolonga-
tion of life in spite of hepatic involvement.

Case Reports

Case 1

This 55-year-old man was admitted on September
29, 1981, with a 4-month history of progressive head-
aches and left-sided weakness. His medical history and
general physical examination were unremarkable. Neu-
rological examination revealed a mild left hemiparesis
with increased deep-tendon reflexes, and a positive left
Babinski response. Skull x-ray films were normal.
Erythrocyte sedimentation rate (ESR) was 82 mm/hr.
Weinberg and Casoni tests were positive. Right carotid
arteriography demonstrated a mass in the posterior
parietal region.

On October 11, 1981, a right parietal craniotomy
was performed. When the dura was opened, the cerebral
convolutions were seen to be strikingly flattened and
widened. A firm multiloculated mass, 7 cm in diameter,
was found located subcortically and was totally re-
moved (Fig. 1). Histological examination of the speci-
men showed areas of marked necrosis. Within the necrotic areas there were numerous communicating vesicles with a lamellar outer layer surrounded by inflammatory infiltration. These findings were consistent with a diagnosis of *E. multilocularis* infestation. The patient’s postoperative course was uneventful. Ultrasonographic examination of the liver disclosed no abnormality, but microscopic examination of a needle biopsy of the liver showed vacuolar degeneration. The patient refused a proposed laparotomy and was discharged with mild hemiparesis on November 5, 1981.

**Case 2**

This 50-year-old man presented with a 5-month his-

![Image](https://via.placeholder.com/150)

ory of dysarthria, inability to identify people he knew, and Jacksonian seizures starting in his right hand. He was hospitalized on December 7, 1981. His medical history and general physical examination, including the abdomen, were unremarkable. Neurological examination revealed him to be confused but able to follow commands. He had papilledema and ankle clonus on both sides. Skull x-ray films were normal. The ESR was 72 mm/hr and eosinophilia was found on peripheral blood smear. Weinberg and Casoni tests were negative. Left carotid arteriography demonstrated a left temporal mass.

At left temporoparietal craniotomy a firm mass was palpable within the temporal lobe. This solid grayish-white multilobulated mass was totally excised. Microscopic examination of the excised specimen was consistent with alveolar hydatid disease (Fig. 2). The patient did well postoperatively. His neurological deficits disappeared except for bilateral ankle clonus. An ultrasonogram of the liver was negative and liver function tests were normal. He was discharged on the 10th postoperative day.

**Comment**

The cerebral alveolar hydatid lesion in this patient was probably primary. No other lesion could be demonstrated. It is interesting to note that this is the only patient with negative Casoni and Weinberg tests.

**Case 3**

This 40-year-old man was admitted on October 17, 1982, complaining of headache of 2 months’ duration and nausea and vomiting in the last 2 weeks. General physical examination revealed moderate hepatomegaly. Bilateral papilledema and a mydriasis of the left pupil...
Cerebral alveolar hydatid disease

were noted on neurological examination. X-ray films of the skull and lungs were normal. The ESR was 86 mm/hr. Casoni and Weinberg tests were positive. An ultrasonogram of the liver showed multiple lesions, and left carotid angiography revealed an avascular left parietal lesion.

At left parietal craniotomy a large firm subcortical mass was removed in toto. Pathological examination of the specimen showed it to be an alveolar hydatid cyst. The postoperative period was uneventful. It was considered that the liver lesions were inoperable due to their number and scattered location throughout the liver. The patient was discharged on November 11, 1982.

Case 4

This 26-year-old man presented on December 7, 1983, with a 6-month history of headache and progressive left-sided hemiparesis. On general physical examination the liver was palpable 15 cm below the right costal margin. Neurological examination showed that the patient was obtunded but arousable, with bilateral papilledema and left-sided hemiparesis. Skull x-ray films were normal. Chest films demonstrated multiple opacities involving both lungs, and an ultrasonogram of the liver revealed a lesion in the right lobe. The ESR was 92 mm/hr. Casoni and Weinberg tests were positive. Right carotid angiography showed an avascular posterior parietal lesion.

At left parietal craniotomy a firm gray subcortical mass was removed totally. Pathological examination revealed cysts of various sizes with an outer layer and surrounding inflammatory reaction, consistent with alveolar hydatid disease. The postoperative course was uneventful. The patient was transferred to the general surgery department, where another surgical procedure was considered to be inadvisable. At follow-up examination 10 months after discharge he was neurologically intact, although the lesions in the lung were larger and the hepatomegaly remained unchanged.

Discussion

The etiological agent of alveolar hydatid disease was first recognized by Virchow as being the larval stage of a cestode of the genus Echinococcus. For a long time, it was distinguished from E. granulosus only by its characteristic hepatic lesions and therefore it was considered a malignant form of E. granulosus. Rausch and Schiller later demonstrated that the alveolar hydatid disease is a morphologically and biologically distinct entity caused by E. multilocularis.

Echinococcus multilocularis, unlike E. granulosus, has a focal distribution. It has been reported to be endemic in Siberia, the Urals, Caucasus, Alaska, St. Lawrence Islands, Canada, Japan, Bavaria, France, Austria, Switzerland, and the northern United States. Cases have been reported from Turkey, Argentina, and Uruguay. Although it has been reported that E. multilocularis is not seen in areas where E. granulosus is endemic, both organisms are endemic in Turkey.

The major definitive host for E. multilocularis is the fox. Dogs and cats can be definitive hosts as well. The life cycle of the cestode includes an intermediate host, lemmings and voles. The ova from the fecal material of the definitive host are ingested by the intermediate host and develop in its liver into vesicular larvae capable of division. The larvae are then transmitted to the fox with its ingestion of the rodent, attach themselves to the duodenum with scolices, and transform into adult taenia of 2 mm.

Man is an intermediate host. The sources of infection of man with E. multilocularis are the infected fecal material from the fox and the dog. Team dogs in northern regions, unboiled thawed water, and skins of hunted fox are important sources of human disease. The principal site of human infection is the liver. The larval cestode exhibits a tumorous, highly invasive proliferation, resulting in vesicular masses and metastasis, particularly to the lungs. Involvement of the brain is rare. A European study of 600 cases of E. alveolaris collected up to the year 1932 showed that in only 31 cases was the brain involved. Brain involvement was primary in only four cases. In a French study in 1977, 81 cases of cerebral alveolar hydatid disease were collected.

The initial lesion of alveolar hydatid disease consists of the larva or the hydatid and the surrounding membranes, the germinative membrane, and an external stratified protective membrane. The latter is not uniformly continuous and arises from the external layers of the germinative membrane. The germinative membrane produces scolices and, by external vesiculation, the secondary cysts. These fertile cysts, which communicate with each other and give the lesion its alveolar appearance, contain scolices and a cytotoxic vesicular fluid. They are surrounded by a zone of fibrotic tissue, and are encircled by epithelioid cells in a palisade arrangement with inflammatory infiltration. Cytotoxic effects of the vesicular fluid result in necrosis of the surrounding structures. The fibrotic and granulomatous tissue surrounding the cysts is not thick enough to impede the passage of these cytotoxic substances, and the necrotic process eventually involves the cysts as well. This development of the parasitic mass results in central necrosis and the formation of a cavity filled with gelatinous magma containing only necrotic tissue. Foci of calcification may appear in the center of these lesions. The area of central necrosis is surrounded by a zone of much smaller young cysts with a thin lamellated outer layer. These cysts are fertile and contain scolices.

The histological diagnosis of alveolar hydatid disease is made from the typical findings of "alveolar" vesicles with a characteristic lamellated outer layer and inflam-
The Russian cases of Chubina and Timofeeva and went surgery for a frontal mass in Tacoma, Washington, and Schiller in 1956, was that of a man who underwent preoperative biopsy revealed scolices. However, review of the pertinent literature shows that these cases are exceptional, and to make the diagnosis the pathologist has to rely on the characteristic histological appearance, rather than on strict demonstration of the parasite. The four patients reported in this paper were male farmers. Their ages ranged from 26 to 55 years. Two of them (Cases 1 and 3) were known to be involved in fox hunting. All of them presented with signs of a cerebral process. In 1978, Thierry, et al., also observed scolices in a case of cerebellar alveolar hydatid disease. Rausch and Martychin reviewed by Thierry, demonstrated scolices in the brain-stem lesions of an autopsied case, but there were no scolices in the older hepatic and pulmonary lesions. These authors were able to find only two previous reports of cases in which scolices were found. Bonis and Sturm also observed scolices in a case of cerebellar alveolar hydatid disease. Smith and Hanson demonstrated "brood capsules" in pulmonary cysts; no scolices were seen. Miguet, et al., in their report of 20 patients with hepatic alveolar hydatid disease, could not demonstrate scolices in any of the specimens examined histologically; on the other hand, immediate examination of specimens obtained from all four patients who underwent preoperative biopsy revealed scolices. Unfortunately, follow-up data are not available in three patients, due to low patient compliance in this region. We have been able to find only five previous cases of surgically treated cerebral alveolar hydatid disease in the literature. The first case, reported by Rausch in 1965, was that of a man who underwent surgery for a frontal mass in Tacoma, Washington. The Russian cases of Chubina and Timofeeva and Martychin, reviewed by Thierry, et al., were also young men. Martychin's case is the only patient operated on for a posterior fossa lesion. Bonis and Sturm in 1969 reported on a 43-year-old Bavarian man with a 7-year history of seizures. In 1978, Thierry, et al., from France reported the only female patient, who was also the only patient operated on for two separate cerebral lesions. Recent reports from the United States and France, as well as this report, indicate a southward progression of *E. multilocularis*.

Conclusions

Our experience with these four cases indicates that cerebral lesions of *E. multilocularis* are amenable to surgical removal, and that surgical therapy provides useful prolongation of life despite the presence of hepatic or pulmonary disease.

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References

Cerebral alveolar hydatid disease


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